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Case Report

Hypoplastic right heart syndrome, absent pulmonary valve, and non-compacted left ventricle in an adult

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ABSTRACT

Hypoplastic right heart syndrome is a rare cyanotic congenital heart disease with under-development of the right ventricle, tricuspid, and pulmonary valves leading to right-to-left shunting of the blood through inter-atrial septal defect. Perinatal mortality is high with very few patients surviving to adulthood without corrective surgery. This report describes a 26-year-old young woman, who had recurrent abortions and stillbirths and detected to have marked cyanosis with hypoplastic right heart, sub-arterial ventricular septal defect, absent pulmonary valve, non-compaction of the left ventricle, and bicuspid aortic valve with aortic regurgitation. The patient died owing to progressive heart failure 4 years after the diagnosis was made.

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1. Introduction

Hypoplastic right heart syndrome (HRHS) is characterized by under-development of the tricuspid and/or pulmonary valves and of the right ventricle (RV) with right to left shunting through an inter-atrial communication.[1] The extreme forms are associated with tricuspid and/or pulmonary atresia. There may be other multiple associated congenital cardiac defects.[2] There are a very few reports of HRHS with patent but severely under-developed right-sided inflow and outflow tracts surviving into adulthood without any repair.[2–4] This report describes a 26-year-old young lady, who was diagnosed to have a cyanotic congenital heart disease (CHD) in childhood but sought medical attention for recurrent abortions and stillbirths in adult life. She was detected to have hypoplastic tricuspid valve and markedly under-developed inlet and trabecular portion of the RV, restrictive subarterial ventricular septal defect, absent pulmonary valve with severely hypoplastic main pulmonary artery, bicuspid aortic valve with moderate aortic regurgitation, markedly dilated but non-compacted left ventricle with ejection fraction of 20% and severe mitral regurgitation.

2. Case report

A 26-year-old woman presented for evaluation of an elevated hemoglobin level of 18 g/dL with normal platelet and white blood cell counts. She had one abortion and two stillbirths in the recent past. The patient had been diagnosed with a heart murmur and cyanosis in childhood; however, her only

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symptom was mild dyspnea on exertion. She had not undergone prior cardiac evaluation. Her physical examination was notable for good physical development (weight 61 kg, height 162 cm, BSA of 1.6), moderate clubbing and cyanosis with an oxygen saturation of 78% on room air, supine blood pressure of 100/80 mmHg, pulse rate of 110/min, engorged jugular veins and suffused face. Cardiac auscultation revealed a quiet precordium, a normal S_1, single S_2, and apical S_3. A grade 3/6 systolic murmur was heard at the apex with a basal early diastolic murmur. Hematologic evaluation revealed a hemoglobin level of 17.8 g/dL, hematocrit of 52%, a white blood cell count of 6.2×10^9/L, and a platelet count of 260×10^9/L. A 12-lead electrocardiogram revealed situs solitus, sinus rhythm, normal atrio-ventricular conduction, and an intra-ventricular conduction defect resembling left bundle branch block. The chest skiagram revealed pulmonary oligemia and cardiomegaly. A transthoracic echocardiogram was obtained for further evaluation of the murmur, hypoxemia, and erythrocytosis (Figs. 1–5). The echocardiogram revealed situs solitus, normally related great vessels, D-loop of the ventricles, and normal systemic and pulmonary venous drainage. The inferior vena cava was dilated with reduced respiratory variation. The right atrium was dilated with inter-atrial septum pushed to the left during inspiration (Fig. 1). The tricuspid annulus measured 14 mm (Z score −2.5); inlet portion of the right ventricular cavity had end-diastolic diameter of 20 mm and the trabecular portion of 12 mm.

There was no moderator band seen in the RV. The sub-pulmonic part of the right ventricular outflow tract was well-developed (28 mm in diastole); the pulmonary valve was absent and virtual pulmonary annulus and the proximal part of the main pulmonary artery measured 14 mm (Z score −2). The tricuspid valve was rudimentary but showed normal motion and mild regurgitation (Fig. 2). The left ventricle was markedly dilated with severely reduced contractile function (ejection fraction 20%). There were prominent trabeculations in the mid and apical segments of the left ventricle with honey-comb appearance and compact ventricular wall was thin (Fig. 4). The aortic valve was bicuspid with moderate aortic regurgitation (Fig. 5). Moderately severe mitral regurgitation was detected on color Doppler interrogation. A small (4 mm) sub-pulmonic ventricular septal defect with left-to-right shunting was noted with a trans-ventricular peak gradient of 70 mmHg (Figs. 3 and 5). Inter-atrial right-to-left shunting was detected on color flow map (Fig. 1). The pulsed wave Doppler examination of the right ventricular outflow tract was notable for pre-systolic and systolic forward flow of markedly reduced velocities and early diastolic retrograde flow (Fig. 3, videos 1 and 2). The tricuspid valve was patent with narrow antegrade color flow jet and a small jet of tricuspid regurgitation (Fig. 6).

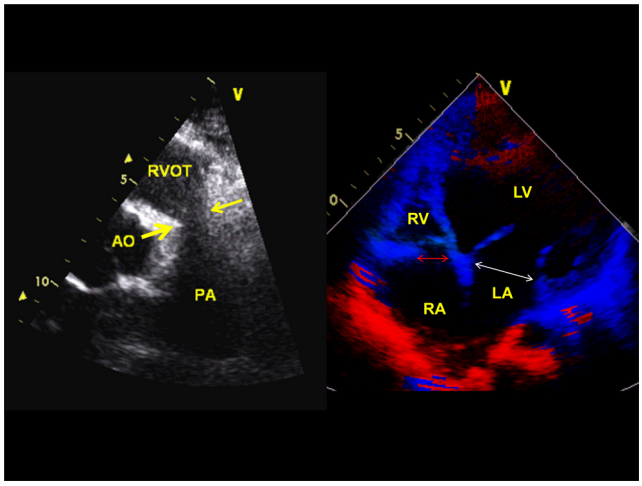

Fig. 2 – Parasternal short-axis view (left panel) showing absent pulmonary valve and hypoplastic pulmonary annulus and the main pulmonary artery. The distal pulmonary arteries are markedly dilated. The right panel shows hypo-plastic tricuspid annulus (14 mm) compared to 29 mm mitral annulus in diastole.

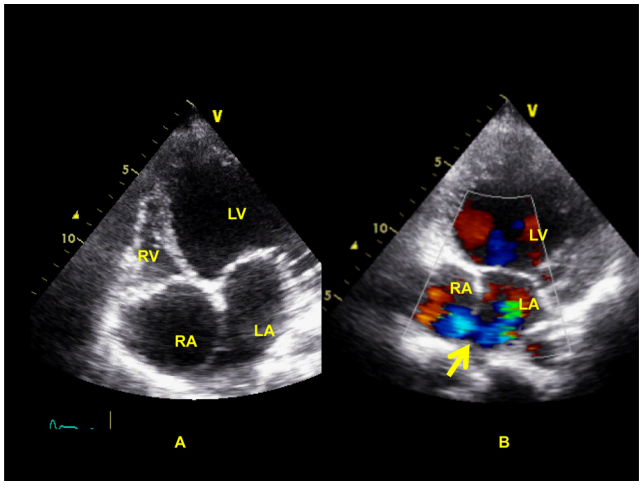

Fig. 1 – Modified apical 4-chamber view on trans-thoracic echocardiographic examination showing dilated right atrium (RA), rudimentary right ventricle (RV), dilated left ventricle (LV) (Panel A) with inter-atrial right-to-left shunting (Panel B, arrows).

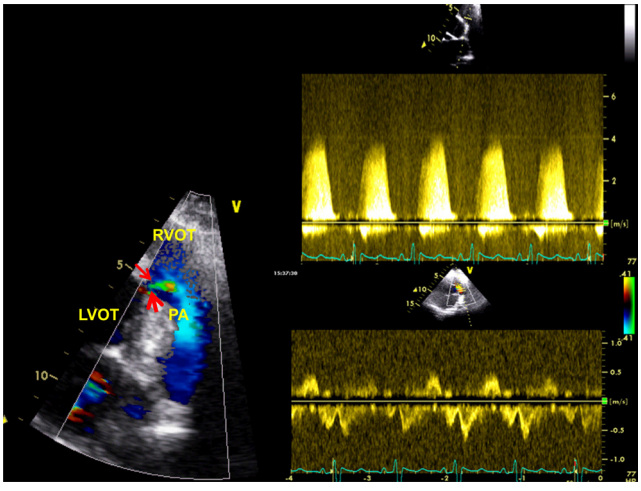

Fig. 3 – Left panel shows small sub-pulmonic ventricular septal defect (red arrows) with a peak trans-ventricular pressure gradient of 70 mmHg (upper right panel). The lower right panel shows low velocity to-and-fro flow across the right ventricular outflow tract.

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